

ANOMALOUS LEFT CORONARY ARTERY ARISING FROM PULMONARY ARTERY

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Origin of the left coronary artery from the pulmonary artery is an uncommon congenital cardiac anomaly. Brooks (1886) first suggested that blood flow in this condition was from the coronary artery into the pulmonary artery. Nevertheless, the view that blood flow is in the opposite direction, i.e. from the pulmonary artery to the coronary artery, was commonly held until recently. Edwards (1958) and others (Abbott, 1927; Agustsson *et al.*, 1962; Baffes, Ketola, and Tatoes, 1961; Case *et al.*, 1958; Lampe and Verheugt, 1960; Sabiston, Neill, and Taussig, 1960) have recently presented evidence supporting Brooks' suggestion.

We are presenting an angiocardiographic and catheter demonstration of flow from an anomalous left coronary artery into the pulmonary artery in a 4-year-old girl.

CASE REPORT

The patient, born in this hospital in 1956, by breech delivery after a normal pregnancy, weighed 2740 g. At the time of her birth, her mother was 32 years old and well, and her father was 38. There were three normal siblings and there had been one miscarriage. At birth, she suffered from asphyxia neonatorum and had a weak cry: oxygen and resuscitative measures were required. The patient developed normally but was noted to be a poor feeder from birth. At the age of 4 months, two weeks before her first admission, she began to wheeze and cough and these symptoms persisted to the time of admission.

On admission, the heart was found to be enlarged to the left with the apical impulse in the 5th to 6th left intercostal space in the anterior-axillary line. A rough, grade III, systolic murmur was heard over the entire præcordium: it was loudest at the apex and was transmitted to the axilla. P_2 was louder than A_2 . Coarse breath sounds and some scattered wheezes were heard bilaterally.

Chest X-ray examination demonstrated a very large heart. Areas of pneumonia were present in both lung apices and the left lower lobe was atelectatic (Fig. 1A).

A cardiogram showed normal sinus rhythm, a mean electrical QRS axis of $+30^\circ$, inverted T waves in leads I, II, aVL, aVF and over the left præcordium, a prominent R wave in the left præcordial leads, and a deep S in V1. The tracing was consistent with severe left ventricular enlargement.

The patient was considered to have congenital heart disease complicated by congestive heart failure and pneumonia. The diagnostic considerations included fibro-elastosis, idiopathic myocarditis, anomalous coronary artery, and glycogen storage disease. There was some immediate improvement with achromycin, penicillin, and digitalis, but after two days of therapy, the infant appeared to be *in extremis*. Chloramphenicol, ACTH, and cortisone were added to try to reverse the apparent rapid deterioration, and there was great improvement: the steroids were discontinued at the end of a week. A chest radiogram taken just before discharge showed clearing of the lungs but no significant change in the heart size or contour.

There were two more admissions during her first year because of failure to thrive and a respiratory infection which precipitated congestive heart failure. During the second of these admissions, a faint diastolic basal murmur was heard. An angiocardiogram demonstrated a huge left atrium and left ventricle with associated poor emptying of the left ventricle, but no shunt was seen (Fig. 1B). The diagnosis most strongly favoured was endocardial fibro-elastosis.

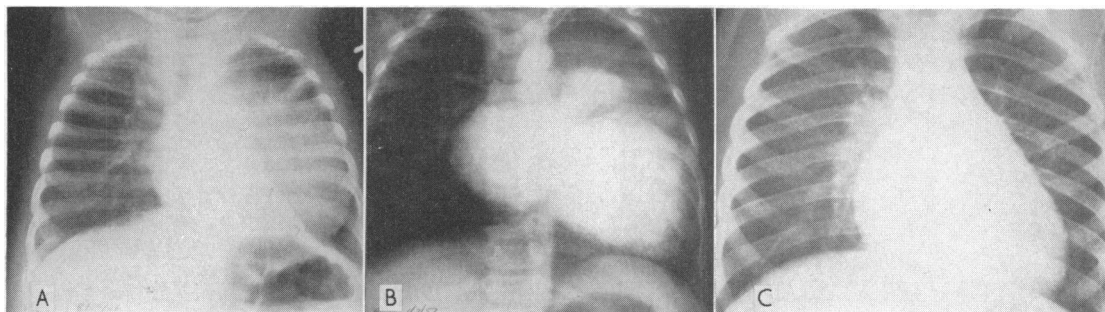


FIG. 1.—(A) Chest radiogram at 4½ months showing great cardiac enlargement to the left, atelectasis of the left lower lobe, and pneumonia in both apices. Note the visible vasculature of right lung is normal. (B) Venous lavo-angiogram at 11 months demonstrates huge left atrium and left ventricle and normal aorta. The coronary arteries were not clearly demonstrated. The right heart was relatively small and no clear evidence of filling of the coronary artery from the pulmonary artery was present on earlier films. (C) Chest radiogram at 4½ years of age, demonstrates normal lungs and some left-sided cardiomegaly but much less than in infancy.

Thereafter, she appeared to be doing better than at any time previously so that at the age of 2½ years, digitalis was discontinued. A cardiogram at 3 years still showed left axis deviation and left ventricular enlargement.

The patient was admitted at age 4½ years for cardiac catheterization and angiocardiology. On examination, no præcordial bulge was demonstrated. The heart sounds were judged to be of good quality. A rough grade II, systolic ejection murmur was heard beneath the right clavicle, over the neck and over the back. No diastolic murmur could be heard. The cardiogram was unchanged from previous examinations. Chest radiogram (Fig. 1C) showed enlargement of the heart which was thought to be mostly left-sided. Laboratory studies were entirely normal. The diagnosis was thought to be aortic stenosis.

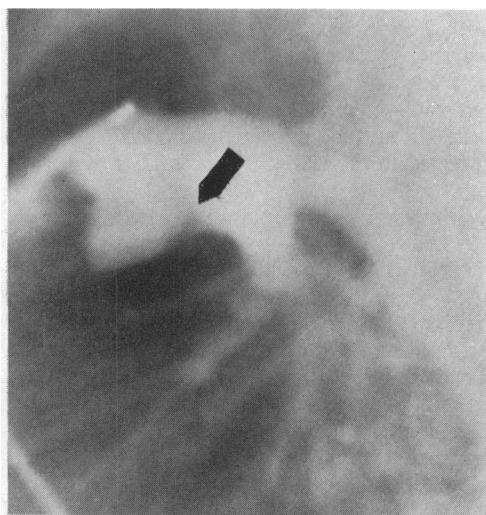


FIG. 2.—Lateral selective angiogram of main pulmonary artery showing persistent filling defect at lower margin of artery (arrow) due to jet of non-opacified blood entering the pulmonary artery via orifice of anomalous left coronary artery.

ANGIOCARDIOGRAPHY AND CARDIAC CATHETERIZATION

A retrograde arterial catheterization of the aorta and left ventricle was carried out but no gradient was found across the aortic valve. A left ventricular angiogram was performed which showed well the large right coronary artery. The left coronary artery filled later than the right coronary artery and, later still, some opacification of the pulmonary artery was seen.

Right heart catheterization demonstrated a significant left-to-right shunt at the pulmonary artery level. The average oxygen saturation of four samples from the right ventricle was 80.1 per cent and of four samples from the pulmonary artery was 84.5 per cent. The difference between these averages is statistically significant ($p < 0.01$) (Grayzel and Jameson, 1963). The pulmonary arterial systolic pressure was slightly raised to 34 mm. while the systemic systolic pressure was normal at 110 mm. Hg. An angiogram performed with the catheter tip in the pulmonary artery showed, on the lateral

views, a persistent filling defect located on the lower border of the image of the pulmonary artery just distal to the valves (Fig. 2). The filling defect represents non-opacified blood entering the main pulmonary artery via the anomalous left coronary artery.

A retrograde aortogram was then performed. Fig. 3 and 4 clearly show, in sequence, the passage of the dye from the ascending aorta into the right coronary artery, through a well-developed anastomotic network into the left coronary artery and thence into the pulmonary artery.

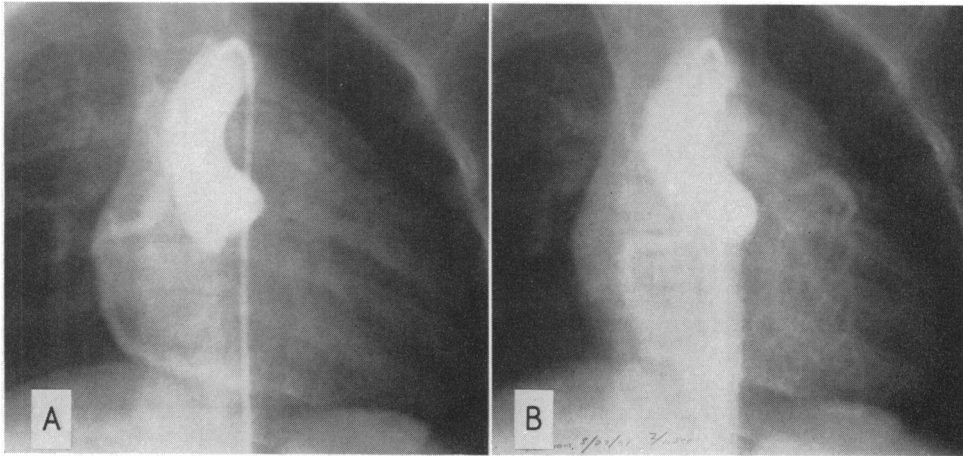


FIG. 3.—(A) Frontal projection of retrograde aortogram at one-third of a second following beginning of the injection, demonstrating immediate filling of a very large right coronary artery and no filling of a left coronary artery. (B) One-third of a second later, a rich anastomotic network carrying opacified blood from the right coronary artery to the anterior descending and circumflex branches of the left coronary artery is well shown.

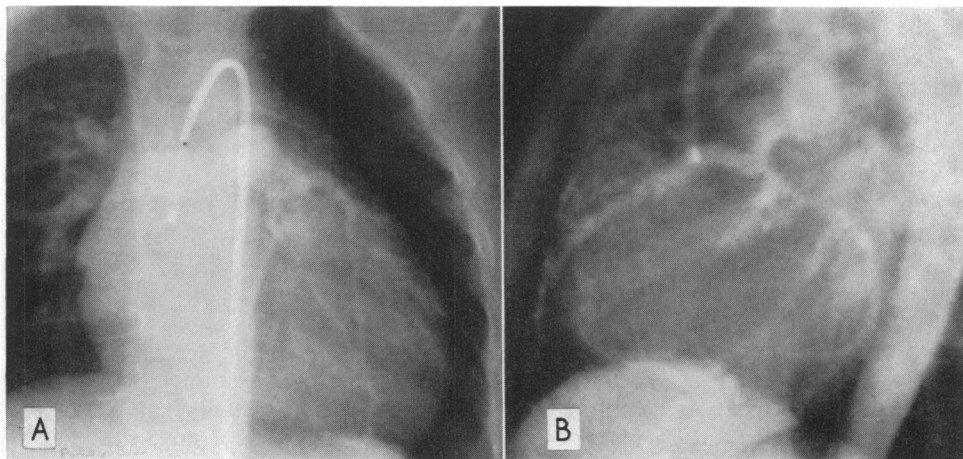


FIG. 4.—(A) 1 second later after the ascending aorta and right coronary artery are cleared of contrast material, the central left coronary artery branches remain opacified as does the pulmonary artery into which blood from the main left coronary artery is flowing in a retrograde fashion. (B) Simultaneous lateral projection showing flow of contrast material from the left coronary artery into the pulmonary artery. Persistent opacification of the anterior descending and circumflex branches and some anastomotic channels is seen.

DISCUSSION

In the case reported above, it has been clearly demonstrated by angiocardiology and supported by cardiac catheterization that in this 4-year-old child, the direction of blood flow is from the anomalous left coronary artery into the pulmonary artery.

The clinical problems presented by patients with this anomaly are best understood with the normal sequence of pressure relations between the aorta and pulmonary artery in mind. Fœtal blood flow is normally from the pulmonary artery into the aorta through the ductus arteriosus. Perfusion pressure of an anomalous left coronary artery under these circumstances is equal to that in the aorta and no disability would be expected. After birth, the pulmonary artery pressure falls as the pulmonary vascular resistance decreases and the ductus arteriosus closes, ultimately reaching an average pressure of about 20 mm. Hg. As the pulmonary artery pressure falls, adequacy of perfusion of the bed of the anomalous left coronary artery is reduced. Another factor affecting the adequacy of perfusion adversely is the desaturation of the pulmonary arterial blood. Presumably about 80 per cent of subjects with this anomaly, after a normal neonatal period, reach a point where perfusion of the left coronary artery vascular bed is so poor that symptoms and signs of myocardial dysfunction ensue. Tachypnoea, respiratory distress with feeding, gross cardiomegaly, and other signs of heart failure characteristically occur, accompanied by clinical and cardiographic evidence of myocardial ischaemia and infarction; and finally death occurs within a few weeks. This critical symptomatic period usually occurs in the second to thirteenth months of life (Fontana and Edwards, 1962; Keith, 1959).

Ideal surgical treatment of these symptomatic infants would be transplantation of the anomalous left coronary artery orifice to the aorta as might be possible using the recently developed techniques of Baffes *et al.* (1961) or end-to-end anastomosis to a systemic artery, but this has not yet been successfully accomplished.

In a few cases, an alternate surgical technique has been successful, i.e. ligation of the anomalous coronary artery (Agustsson *et al.*, 1962; Case *et al.*, 1958; Sabiston *et al.*, 1960). To be successful, the procedure must improve coronary perfusion, and logically this could only result if predominant blood flow was from the anomalous left coronary artery into the pulmonary artery as observed in Sabiston *et al.*'s case (1960).

In those instances in which ligation resulted in death, flow was presumably from the pulmonary artery into the left coronary artery so that ligation, rather than improving coronary perfusion, reduced it. Talner, Stern, and Figley (1958) have demonstrated blood flow from the pulmonary artery into the anomalous coronary artery in two infants. We, too, have demonstrated this direction of flow in a 10-week-old infant in whom the diagnosis was later proved at necropsy (Ellis and Jameson, unpublished data). In such infants, ligation of the coronary artery could not be expected to be successful except possibly in those in whom the direction of flow may have been momentarily reversed by the angiocardiology injection resulting in a false picture of flow from the pulmonary artery into the anomalous left coronary artery. To avoid this latter possibility, all patients in whom the diagnosis has been made might be tested for a left-to-right shunt at the level of the pulmonary artery using such a sensitive method as Clark and Barger's (1959) platinum-hydrogen electrode, thus providing a rational basis for selecting cases for treatment by ligation of the anomalous coronary artery.

Some subjects with this anomaly survive to adult life, and they are characteristically asymptomatic until their usually unexpected and sudden death. Baylis and Campbell (1952) reported the case of a woman who died aged 76 from recurrence of carcinoma of the breast: she had enjoyed good health till terminally. Spontaneous long-term survivals can only result when anastomotic interarterial (intercoronary) communications develop sufficiently to maintain adequate perfusion of the left coronary artery vascular bed. Since adequate left coronary artery perfusion pressure exceeds that normally present in the pulmonary artery, the anastomotic circulation must be sufficient to supply a large enough flow of blood from the left coronary artery into the pulmonary artery to maintain a significant pressure gradient between the peripheral components of the left

coronary artery vascular bed and the pulmonary artery. That blood flow is from the anomalous left coronary artery into the pulmonary artery in long-term survivors is supported by evidence cited by Edwards (1958), Jurishica (1957), George and Knowlan (1959), and Lampe and Verheugt (1960), and most recently in 2 of the 3 cases reported by Agustsson *et al.* (1962). The surviving adults have usually not had a known history of illness in infancy, although the reliability of the evidence may be questioned.

Agustsson *et al.*, in their excellent presentation of three cases of this anomaly (1962), indicate their belief that such cases fall into either of two groups—an infantile type with inadequate inter-coronary anastomoses and an adult type with adequate anastomoses. They imply that the group into which a given case falls is determined “by basic differences in the coronary circulation present at birth, rather than to consequently developed collateral circulation in the latter” [adult type]. Of particular interest, therefore, is the child here presented, who was closely followed during a long period of severe cardiac disability beginning at about the age of 5 months with manifestations of the infantile type. Presumably, during this period, left coronary perfusion was barely adequate to support life. In contrast, at 4 years of age, she had a substantial shunt from the left coronary artery into the pulmonary artery with a continuous murmur, more modest X-ray and cardiographic evidence of left ventricular enlargement, and she was asymptomatic—typical manifestations of the adult type. Obviously, then, it is possible for some subjects presenting in infancy with manifestations of severe myocardial insufficiency due to this anomaly in its infantile form to develop sufficient intercoronary anastomoses to become typical asymptomatic cases of the adult type in later life.

Whether a specific symptomatic infant with this anomaly will survive must depend on numerous factors. Support of the left coronary artery perfusion pressure by a very gradual, as opposed to sudden, fall in pulmonary artery pressure during the early months of infancy must be of importance.

In our patient the possible supportive effect on left coronary artery perfusion of a very prolonged course of congestive heart failure as well as of the short course of cortisone are interesting questions. Although this child now has a substantial shunt between the left coronary artery and the pulmonary artery and since, therefore, either ligation of the left coronary artery or its anastomosis to a systemic vessel would theoretically improve left coronary perfusion pressures, we are not yet certain of the proper therapy to be offered as the patient is now asymptomatic.

SUMMARY

A case of anomalous left coronary artery originating from the pulmonary artery, with unequivocal angiocardiographic proof of blood flow from the aorta to the right coronary artery, thence through an anastomotic network to the left coronary artery and finally into the pulmonary artery has been presented. The clinical picture of such patients at different ages has been outlined in relation to the nature and extent of the anastomoses existing between the coronaries and the direction of flow in the left coronary artery. Some of the problems in the choice of a therapeutic approach have been noted.

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